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ON THE ROTATIONAL DEFORMITY OF THE SHOULDER FOLLOWING AN OBSTETRIC BRACHIAL PLEXUS INJURY

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To Ursula

ABSTRACT

An internal rotation deformity of the shoulder occurs very frequently in brachial plexus birth palsy. Even though surprisingly accurate descriptions of the deformity were already published at the beginning of the 1900s, the nature of the deformity is not well understood and there is no consensus regarding surgical treatment. This thesis was aimed at improving the scientific basis for surgical treatment of the deformity.

In study I the passive mechanical properties of single cells and muscle bundles were investigated in muscle biopsy specimens harvested from the subscapularis muscle in nine children with birth palsy who had undergone open surgery for shoulder contracture. Biopsy specimens from seven healthy subjects were used as controls. Single muscle fibres from patients with birth palsy displayed a shorter slack sarcomere length and linear deformation of the fibre within a wider zone of sarcomere length. There was a greater relative increase in stiffness for fibre bundles / single fibres compared to the controls.

Study II was an investigation of the histopathology of muscle biopsies harvested from the subscapularis muscle in 13 children operated on for brachial plexus birth palsy. The majority of the subscapularis muscle biopsy samples had an essentially normal morphology and showed a predominance of type I myosin heavy chain isoform (slow fibres), while one sample showed signs of fibrosis. The findings of studies I and II are interpreted to be consistent with the theory that the shortening of the subscapularis in a majority of patients is a result of altered muscular balance following the nerve injury.

Study III was an evaluation of the results at one year after surgical correction of internal rotational deformities in the shoulders of 270 patients with birth palsy, using open subscapularis elongation and latissimus dorsi-to-infraspinatus transfer, alone or in combination. Open relocation was performed on incongruent joints. Ninety-two per cent (97 of 105) of the incongruent joint could be relocated, with an upper age limit of 12 years for the subluxed joints and 5 years for dislocated joints. There was a substantial overall mean improvement in external rotation and the Mallet score following surgery and a notable decrease in the mean internal rotation for the relocated joints, but not for the congruent joints. The trumpet sign was corrected in 83%. Adding a latissimus dorsi transfer did not result in greater improvement in the mean external rotation compared with elongation of the subscapularis alone.

Study IV was an evaluation of the long-term effects in 118 of the patients from study III, using the same protocol. There was an overall moderate decrease in the mean rotational range compared with the results at one year post surgery, while abduction remained unchanged, as did the correction of the trumpet sign. One out of four patients with relocated joints had required additional surgery in order to adjust the range of rotation or to stabilise the joint.

In conclusion, the open subscapularis elongation produced a good long-term correction of the deformity with lasting elimination of the trumpet sign and reduction of incongruent joints and with a moderate mean loss of internal rotation. Long-term monitoring of the patients operated on is recommended because of the large individual variations and the need for additional surgery in selected patients.

LIST OF PUBLICATIONS

This thesis is based on the following papers, which are referred to in the text by their Roman numerals:

I. Subscapularis muscle mechanics in children with obstetric brachial plexus palsy. Einarsson F, Hultgren T, Ljung B O, Runesson E, Friden J. J Hand Surg Eur Vol, 2008; 33 (4): 507-12.

II. Structural characteristics of the subscapularis muscle in children with medial rotation contracture of the shoulder after obstetric brachial plexus injury. Hultgren T, Einarsson F, Runesson E, Hemlin C, Friden J, Ljung B O. J Hand Surg Eur Vol, 2010; 35E (1): 23-28.

III. Surgical correction of a rotational deformity of the shoulder in obstetric brachial plexus palsy. Short-term results in 270 patients. Hultgren T, Jönsson K, Pettersson H, Hammarberg H. Bone Joint J 2013;95-B:1432-8

IV. Surgical correction of the shoulder rotation deformity in obstetric brachial plexus palsy. Long-term results in 118 patients. Hultgren T, Jönsson K, Roos F, Pettersson H, Hammarberg H. Manuscript.

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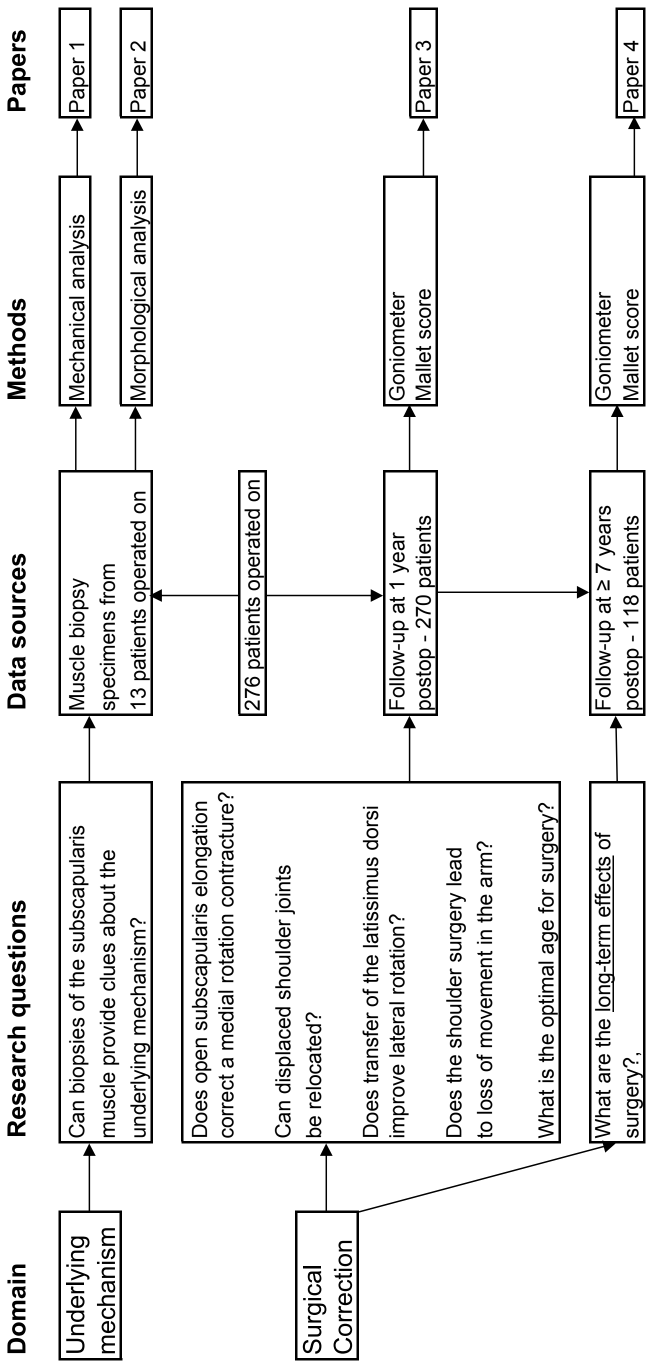

ABBREVIATIONS

ADL	Activities of daily living
AMS	Active Movement Scale
BPOM	Brachial Plexus Outcome Measure
CI	Confidence interval
CT	Computerised tomography
HE	Haematoxylin-Eosin
HTX/eo	Haematoxylin-Eosin
He-Ne	Helium-Neon
MHC	Myosin heavy chain
MRI	Magnetic resonance imaging
MRC	Medical Research Council
OBPP	Obstetric brachial plexus palsy
SEM	Standard error of the mean
SL	Sarcomere length
ROM	Range of motion

INTRODUCTION

Injuries to the brachial plexus can have a considerable impact on the function of the upper limb. Fortunately these injuries are rare. The obstetric plexus injury in Sweden has been reported to occur in 1.4 to 2.7 per 1000 live births.[1-3]

The cause of the birth injury has been attributed to various intrauterine[3-5] and extrauterine[6-8] factors. The widely agreed upon mechanism is a combined traction and lateral pressure on the head during the late stages of a difficult delivery where the shoulders are stuck in the birth canal. This traction can be severe enough to cause partial or complete ruptures of the nerves in the plexus area. In a prospective population based study of over 30000 vaginal deliveries Mollberg et al. demonstrated that births which resulted in brachial plexus injuries with persistent motor impairment were all characterised by substantial difficulties in delivering the shoulders and greater force had been applied than in the group which had only transient motor symptoms.[9]

Most frequently, the injury is limited to the upper spinal nerves and these patients have a considerable potential for spontaneous healing.[1,10,11] The designation 'Erb-Duchenne palsy' or 'Erb's palsy' refers to a C5-C6 injury, with subsequent paralysis of the shoulder and of elbow flexion. If the wrist and finger extensors are also paralysed this may indicate that C7 is also involved. The majority of these partial plexus injuries are benign in nature and probably 70–80% recover spontaneously.[2,12] If the palsy does not resolve completely within 2–3 months, there will usually be residual weakness to a lesser or greater extent.[1,13-16] Figure 1 shows a child of 3 months with the typical arm posture of an Erb's palsy.

Plexus injuries involving the upper as well as the lower parts of the plexus constitute a very different situation. Not only is the whole limb including the hand affected, the tearing of the nerves is usually more complete and the prognosis is much poorer. Most of these patients benefit from nerve reconstruction. Fortunately, this type of injury is rare; it represents approximately 10% of the children with a persistent palsy.[1,2,14,17]
Breech deliveries can cause plexus injuries with a slightly different pattern. They are usually limited to C5 and C6, but they are often bilateral, and the spinal nerves are injured at a more proximal level, even torn right out of the spinal cord (root avulsion). The

clinical picture in these children is characterised by extremely atrophic and atonic shoulder muscles and elbow flexors.[18]

Considering that the forces of the birth trauma are strong enough to rupture or avulse several spinal nerves, and occasionally also fracture bones in the limb, it is reasonable to assume that there may also be substantial injuries to other surrounding tissues, such as tendons, muscles and joints. Not much is known about the associated soft tissue injuries, however.

Fig. 1. A young boy with Erb's palsy, showing the characteristic position of the arm**.** There are signs of weakness in the wrist extensors in this photo but this patient was clinically labelled as having a C5-C6 injury.

BACKGROUND

HISTORICAL NOTES

Surgical treatment of obstetric plexus lesions began in the early 20^{th} century, with the first recorded nerve repairs by Kennedy in 1902[19] and one year later by Taylor.[20] The early nerve repairs relied on direct suture rather than nerve grafts, and the results were not convincing.[10] The surgical repair of the birth lesions gradually disappeared from the scene and would not reappear until the late 1970s, with the advent of safe anaesthesia, microsurgical techniques and the use of autologous nerve grafts and through the pioneering work of Alain Gilbert.[21]

In parallel with the early attempts at nerve repair, surgeons recognised that the children with partially healed birth injuries developed characteristic deformities, particularly in the shoulders and elbows and thus began the pursuit of an alternative path of surgical treatment. These procedures are orthopaedic in nature and are often referred to as secondary surgery, because the aim is to improve the function in a limb with manifest deformities after a partially recovered nerve lesion. Release of tendons and muscles in order to improve the mobility were introduced at the beginning of the 1900s.[10,22] Muscle transfers to enhance the power of selected movements were introduced in the 1930s.[23] Osteotomy was used early on to adjust the available range of motion of the limb to a favourable sector.[24] Posterior subluxation and dislocation of shoulder joint following obstetric plexus lesions was described as early as 1905 by Whitman[25], who proposed treatment by closed reduction in the anesthetised child, using repeated stretching and relaxing with slowly increasing force. Fairbank[22] commented in 1913 that the joint deformity ‘seems to be extremely common’ and he advocated an open anterior approach with division of the subscapularis tendon, the joint capsule and the supraspinatus tendon. The arm was then placed in a brace in abduction and external rotation for three months. Sever[10] used a similar approach but also divided the insertion of the pectoralis major and the conjoined tendon, with the tip of the coracoid. Sever was adamant in his view that the joint capsule should be left intact. These early reports provide very little detail regarding the postoperative results.

THE ROTATIONAL DEFORMITY

An internally rotated position of the arm can be seen shortly after birth in nearly all children with a plexus injury. The shoulder muscles are paralysed, except for the pectoralis major, which usually retains some power due to its broad innervation from many levels in the brachial plexus and this sets the arm in internal rotation. Thus an increasingly powerful internal rotator is usually present from the beginning, while the most important and less powerful external rotators; the infraspinatus and teres minor may take months or years to recover. A contracture in the internally rotated position may develop at any time during the first decade, but it is most frequently manifested within the first two years. The rotational contracture is very common; it has been reported to occur in 45% of children with birth palsy who did not have full neurological recovery by three weeks.[15]

The rotational deformity limits the ability to reach with the hand anteriorly, laterally and cranially. The most obvious sign in a patient with a severe limitation of external rotation is the so-called trumpet sign, in which the elbow is elevated as the hand is brought to the mouth (Figs. 2 & 4a). A rotation deformity can also make it impossible to reach with the hand to the back of the neck (Fig. 3). The ability to work with the hand on a desk top can be severely impaired (Fig. 4b)

Fig. 2. Hand-to-mouth**.** A boy with Erb's palsy and excellent spontaneous recovery from the nerve injury but with a prominent trumpet sign.

Fig. 3. Hand-to-head. A teenager with Erb's palsy, unable to reach the back of the neck because of an internal rotation deformity.

Fig. 4 a. A lady of 70 years with Erb's Palsy and a marked trumpet sign.

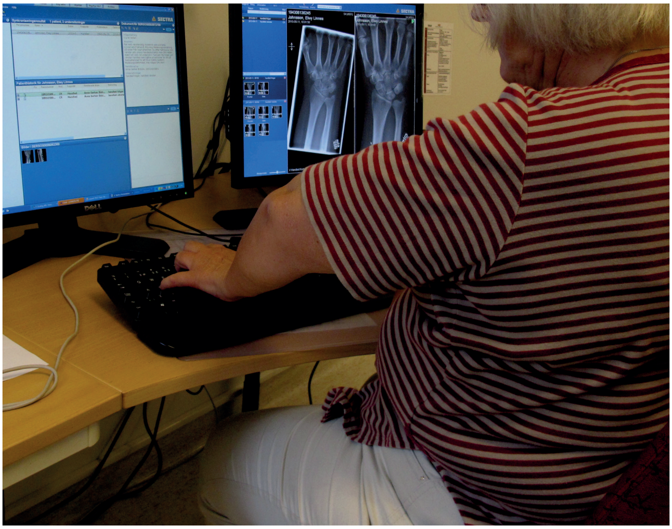

Fig. 4b. The same patient as in Fig. 4a. She was unable to work on a keyboard with the affected limb because of the rotation deformity of the shoulder.

SURGICAL TREATMENT OF THE ROTATIONAL DEFORMITY

Nerve reconstruction of the birth injury is a well-established and uncontroversial procedure, particularly for the injuries involving the entire plexus. The indication is more limited in the partial, Erb's type lesions and a nerve reconstruction can neither fully restore function nor prevent the development of the characteristic deformities.[11,26-28]

Thus the need for effective secondary procedures is likely to remain, despite advances in the surgical repair of the plexus lesion. The most serious sequel in the partial lesions is the internal rotational deformity of the shoulder. Paradoxically it is most common in cases where the global function, including shoulder elevation, is very good and where there is no indication for nerve repair.[28-30]

Three components need to be considered when dealing surgically with the rotation deformity;

- Contracture in internal rotation
- Weakness in external rotation
- Joint deformity

The contracture

There is a convincing documentation in the literature to support the view that a shortening of the subscapularis is the main ingredient in the rotation contracture.[22,28,31,32] The first surgical interventions relied on open tenotomy of the subscapularis[22], and often other muscles were divided as well.[10,33] An alternative treatment, which preserves the integrity of the subscapularis tendon and the joint capsule is the 'subscapularis slide', where the origin of the muscle is detached from the scapula and it is allowed to slide laterally.[31] Good results have been reported with this technique.[31,34,35] Gilbert reported a recurrence rate of 18% when the procedure was done after the age of two years.[35] A recent study reported that 50% of the contractures recurred when the procedure was used alone without muscle transfer.[36]

In 1996 Birch and Chen[37] described a modification of the early Fairbank procedure; instead of just sectioning the subscapularis tendon it was divided in a step-cut fashion and then carefully repaired in elongation. Later Pearl et al.[32] described arthroscopic release with capsulotomy and tenotomy. The latter two techniques have been reported to provide good release of the internal contracture, although some loss of internal rotation has also been reported with both techniques.[32,38]

The weakness in external rotation

L'Episcopo[39] remarked in 1939 that although the release of tight internal rotators, as advocated by Sever,[10] was very satisfactory, the deformity tended to recur. He therefore combined tendon releases with muscle transfers to augment the power of external rotation. The insertions of both the latissimus dorsi and the teres major were detached, rerouted around the outside of the humerus, and then attached to the upper end of the humerus, just above the origin of the lateral head of the triceps. Hoffer[40] suggested a modification, in which the two tendons are attached to the rotator cuff. Variations of these techniques have gained widespread popularity. Most authors today use only the latissimus dorsi for the transfer. The published results are good and some authors report that the procedure improves external rotation as well as abduction.[35,41] In the majority of reports a combination of muscle release and muscle transfer has been used, which makes it difficult to evaluate the effect of the transfer *per se*.[32,41-43]

The joint deformity – classifications and surgical procedures

After the early descriptions of subluxation and dislocation at the beginning of the 1900s, interest in the joint deformity appears to have faded.[10,22,25] In 1955 Wickstrom et al.[44] described open reduction and transfixation of the joint with Steinmann pins for four weeks, with good results in four out of five patients operated upon. Zancolli,[45] in 1981, commented that posterior subluxation was most likely a combined effect of joint trauma and altered muscular balance, and that the only rational treatment was to do an externally rotating osteotomy of the humerus in order to restore some external rotation of the arm.

In the late 1980s Birch began a systematic clinical study of the shoulder joint deformities in children with birth palsy.[28,37] He described four types of deformity: simple and complex subluxation and simple and complex dislocation.[46] The classification was based on clinical examination and standard radiographs, with the term 'complex' indicating associated deformities of the coracoid, acromion and glenoid. In a series of 183 patients with subluxation and dislocations published in 2006, successful relocation was achieved in 163, with a mean gain in external rotation of 58° and a mean loss of internal rotation of 10°.[28]

In 1998 Pearl et al.[47] published a classification based on intraoperative arthrograms of 25 children with rotational contractures. These authors identified three types of deformity; (subluxation with) flattening of the posterior glenoid, biconcave glenoid with the humeral head resting in the posterior facet and 'pseudoglenoid' with the humeral head 'resting in a distinct, retroverted posterior articular surface.' Pearl later reported MRI-verified remodelling of the glenohumeral joints in 11 of 15 children who had been operated on with arthroscopic release of the subscapularis tendon and the anterior joint capsule and relocation of the joint.[32]

With the widespread use of CT and MRI, new classifications have emerged. The Waters classification,[48] uses a combination of glenoid version, as measured on tomograms according to Friedman[49] and the degree of posterior shift of the humeral head, also measured radiologically. In 2001 van der Sluijs et al.[50] presented a series of 16 children younger than 12 months with unresolved birth palsy who had been examined using MRI. A qualitative classification with three types of glenoid was proposed; concave-flat (normal), rounded and biconcave. These authors could not differentiate between the classes of biconcave and pseudoglenoid described by Pearl et al.[47]

In recent years glenoplasty[51] and anteversion osteotomy[43] have been introduced as means to deal with severely deformed joints, where relocation is not possible. A posterior positive wedge osteotomy with bone graft is used to correct the retroversion of the glenoid fossa, thus providing a stable support for the humeral head. Early results are promising.

The long-term development of relocated joints and how the development and remodelling are influenced by the age at joint relocation is not well known.

With so many surgical procedures and combinations of procedures available, and with the very limited and heterogeneous study populations at hand, the scientific evaluation of surgical treatment becomes a great challenge.

THEORIES ON THE CONTRACTURE MECHANISM

If there is overwhelming documentation to support that a shortening of the subscapularis is the main ingredient in the rotation contracture, the mechanism behind this muscle shortening is not well understood. Two theories have dominated in the literature.

The majority of authors subscribe to the theory that a lack of muscular balance is the main cause; i.e. the nerve injury results in an early dominance of internal rotators and a lack of power in their antagonists. When the external rotators recover (which may take two years or more), the contracture is already manifest.[23,35,50,52,53]

The second theory speculates that the birth trauma may cause a direct injury to the muscle, leading to fibrosis and shortening. This has been proposed by Zancolli.[54] In their report on a series of 183 operated patients with rotation contracture and incongruent shoulder joints, Kambhampati et al.[28] noted signs of fibrosis in 42 of the subscapularis muscles, although the fibrosis was not verified histologically.

MRI studies on the muscles around the shoulder have shown that the muscle atrophy is most prominent in the subscapularis.[55-57] Waters et al.[57] demonstrated that if the pectoralis major was included as an internal rotator, the combined cross-sectional area of this muscle and the subscapularis was greater than the combined area of the external rotators. They found that an increase in the radiologically measured imbalance correlated with the degree of joint deformity but did not correlate with reduced joint mobility as measured by the Mallet score.

In recent years a third theory has emerged, which suggests that a neonatal injury at the plexus level causes impaired growth of the denervated muscles, with subsequent muscle shortening and joint contracture. Experimental studies on mice have demonstrated such a mechanism.[58,59] Following experimentally created plexus injuries in neonate mice, impaired longitudinal growth (and corresponding contracture) was verified in the biceps and subscapularis. The reduction in longitudinal growth was inversely proportional to the degree of motor recovery; thus the more complete the denervation, the more severe the shortening and the contracture.

EVALUATION OF THE RESULTS OF SURGERY

This is fundamental in dealing with any surgical treatment. Without reliable and valid outcome measures we cannot know what we achieve (or don't achieve) with our surgical interventions.

Two areas are of special concern in the evaluation of secondary surgical procedures in patients with obstetric brachial plexus palsy (OBPP):

- How do we evaluate and compare results over time in developing children?
- How do we measure mobility in the smallest children?

The time factor

Patients with OBPP are a limited and heterogeneous study population which makes it very difficult to conduct controlled studies. It might be possible to randomise between two surgical procedures that we regard as more or less equal. A comparison between a treatment group and a non-treatment group would raise serious ethical issues. That leaves the option of doing uncontrolled longitudinal studies.

In general before and after studies have a weaker evidential value than studies with control groups because there is a problem with causality. It is difficult to separate the possible effects of time-related factors from those of the interventions. This is a particular concern when dealing with the OBPP patients, for two reasons:

The healing time for an Erb´s type injury can be two years or more, during which time the motor function in the shoulder and elbow can be expected to improve.

Children will perform better over time as they mature and become more adept in various motor functions.

Once the healing of the nerves is complete, it is reasonable to expect that we can measure and compare ROM accurately over time, with careful and consistent examination techniques. The measurements that we register in the smallest children should be regarded as less reliable than in the older children.

We can gain a certain degree of control of the time-related influence by dividing the study population into separate age groups and also by comparing the short-tem results of surgery with the long-term results.

Comparing *function* over time may be impossible. A few years of difference in age can have a profound effect in a young developing child. Even a fairly simple semi-functional test such as the Mallet score can be difficult for the smallest children, but easy for a five-year-old.

Joint mobility

Range of motion (ROM) is technically easy to record, even in young children, provided that we have standardised reference lines and sighting lines, and provided that we consistently use a goniometer.

The importance of standardised reference lines becomes very evident in measurements of shoulder mobility. Patients with impaired function will inevitably compensate for a lack of elevation by arching the spine. In such a case an imagined tangent through the upper trunk should be used as a reference line towards the humerus (Fig. 5).

Flexion is measured with the patient viewed from the side, while abduction is easiest to measure from the back, with an imaginary tangent through the upper thoracic spine as reference.

A lack of shoulder rotation will be compensated for by rotating the trunk and to some extent by moving the scapula in the anteroposterior plane. Using an imaginary tangent through both shoulder joints, as viewed from above, with the arm relaxed in adduction, and the elbow in 90° of flexion will give a reference line towards the forearm. The sighting line is parallel with the humerus. When testing internal rotation the front of the trunk will limit the motion. This can be avoided by supporting the elbow from behind in just enough shoulder flexion to allow full internal rotation of the arm.

The techniques described here measure the sum of glenohumeral and thoracoscapular movement. Measuring the active glenohumeral movement in small children isn’t possible in the clinical setting, although in external rotation the scapula automatically has a degree of stabilisation against the thorax, which helps to isolate the humeroscapular motion.

A specific problem with children (and some adults) is that they may or may not cooperate and we cannot be sure that they will make a maximum effort, particularly if we need to do many measurements in a row. The examinations must be performed in a playful and relaxed atmosphere. A friendly and unassuming interaction with the parent(s) goes a long way towards putting the child at ease, as does a good supply of interesting toys and plenty of time.

With the smallest children it is often easiest to evaluate specific movements by letting them reach for desirable toys that are held in relevant positions (Figs 5 & 6), while a second person takes measurements with the goniometer. Some children respond well to a game of imitating the movements of the examiner and many enjoy a good challenge: ‘Can you do better than me, him, her, that…’

Fig. 5. A child arches the back to assist in elevation of the arm. Approximate reference lines for measurement of shoulder flexion have been superimposed on the photo.

Fig. 6. Testing abduction in a child of 9 months

The Active Movement Scale (AMS) is an ordinal eight-grade scale that was developed and validated in Toronto.[60,61] The AMS is particularly useful as an instrument for evaluating limb function in the smallest children, in order to help in the selection of candidates for nerve reconstruction and for evaluation of the results of the surgery. It can be used for selected movements or as an aggregate score. An advantage of the AMS is that it can be used as an evaluation tool during the entire life of the patient. A disadvantage of this instrument in the context of the shoulder rotational deformities is that it does not have quite the sensitivity for the specific movements that we need to study in this subcategory of OBPP patients.

The Mallet score. The French neurologist Mallet introduced the most widely used scoring test for global shoulder function in OBPP.[62] It is a semi-functional test, which comprises two pure shoulder movements; abduction and external rotation, and three functional composite movements which involve shoulder rotation: hand-to-mouth (Fig. 2), hand-to-head (Fig. 3) and hand-to-back.

Each movement is classified in three or five grades, depending on which system is used,[62,63] so that the aggregate score can vary from 5 to 15 or from 5 to 25.

The Mallet test has been shown to have intraobserver and interobserver reliability[64] and it is used by all who examine children with birth palsy.

Perhaps the greatest value of the Mallet test in the context of the rotational deformities of the shoulder is that it provides a grading of hand-to-mouth (trumpet sign) and of hand-to-head (reaching the back of the neck). Both of these functions are highly dependent on external shoulder rotation and impairment of either reflects problems in activities of daily living (ADL) related to the internal rotational deformity.

A disadvantage of the Mallet test, as with all sum scores, is that the sum does not tell us which specific function is affected. A reduction of, e.g., 4 points in the aggregate score can indicate problems in shoulder elevation and/or shoulder rotation and/or elbow flexion. Furthermore, since it is an ordinal scale, calculations using means of scores shouldn't be used, although in actual practice this is done by most authors. One needs to be aware of the fact that a change in mean score, even for a separate movement, can have different interpretations. For example, a reduction in the mean score for hand-to-back, could mean that a number of patients have gone from 'easy' to 'difficult,' which is not so bad. It could just as well mean that patients have gone from 'easy' and 'difficult' to 'impossible,' which is more of a problem.

The trumpet sign, although characteristic of a internal rotational deformity, can also be the result of other motor impairments. Weakness in the elbow flexors is often compensated for by raising the elbow in the air, thus eliminating gravity. A supination deformity may be compensated for by rotating the shoulder internally, which may mimic a true trumpet sign.

Function

Our efforts to help these children do not stop at increasing the mobility of specified joints. What we aim for is improved function in daily activities. Very little has been published on how the secondary surgical procedures in OBPP patients affect their ADL.

In part this is because of the lack of control groups and the difficulties in comparing the effects of surgery over time in developing children. Before and after studies have limitations. How do you determine if surgery at the age of two leads to improved function when the children operated on are ten, without a control group?

Another problem is that we need to identify and define which functions and activities that are affected by the rotational deformity of the shoulder. What is the functional gain for the patient when we restore the external rotation and how is the function affected by a reduction of the internal rotation?

A recent study by Westin et al.[65], based on interviews, looked at the short-term effects of surgical correction of the shoulder rotational deformity in a group of children and adolescents with OBPP. This study showed a high level of satisfaction with the results of the surgery, particularly with respect to eating with a knife and fork, drinking from a glass, holding a handset to the ear and with respect to general mobility. There was also a high level of satisfaction with the cosmetic improvement, whereas the levels were lower for taking objects from a shelf at head height and for improvement of strength. In another recent study healthy adults were tested in daily activities while wearing a brace which artificially limited external rotation of the shoulder.[66] The activities which were most affected by the limited rotation were eating with spoon, pouring water from a pitcher, combing the hair and drinking from a glass, corroborating some of the findings in the study by Westin et al. More studies are needed in this area.

The Brachial Plexus Outcome Measure (BPOM)[67] is a recently developed disease-specific assessment tool that should help considerably in evaluating the effects of secondary surgical procedures in OBPP patients. It is used to grade a total of eight activities which have been selected to assess function at three levels of the upper limb: shoulder, elbow/forearm and wrist/hand.

OVERRIDING AIM OF THE THESIS

To provide scientific support for an algorithm for the surgical treatment of the shoulder rotation deformity in obstetric plexus palsy.

The project addresses the following domains:

- What are the mechanisms behind the rotation deformity?
- What are the immediate results of surgery?
- What are the long-term effects of surgery?

Research questions:

- Can biopsy specimens from affected subscapularis muscles provide clues about the underlying mechanism for the deformity?
- Does open subscapularis elongation improve external rotation?
- Does transfer of the latissimus dorsi muscle contribute to the range of external rotation?
- Can a displaced shoulder joint be relocated?
- How are the results of surgery affected by deformities of the joint?
- Are there age limits for successful surgery?
- Does the surgery lead to loss of internal rotation or other movements in the arm?
- What are the long-term effects of surgery on mobility and joint stability?
- Is there a need for additional surgery over time?

AIMS OF THE STUDIES

Study I

The aim of the study was to analyse the passive mechanical characteristics of muscle fibres from the subscapularis in children with OBPP and rotational contracture, in order to identify and evaluate possible changes in the muscles, specific to this condition.

Study II

The aim of this study was to see if we could identify morphological changes in biopsy specimens from the subscapularis muscle, which might explain the nature of the muscle shortening in patients with birth palsy and shoulder rotation contracture.

Study III

The aim of this study was to evaluate the short-term results of open subscapularis elongation, with or without latissimus transfer, for correction of the shoulder rotational deformity in patients with OBPP. The primary aim was to evaluate the mean changes in joint mobility and the Mallet score from pre- to post-surgery and possible interactions with respect to whether or not a muscle transfer was used, to the condition of the joint, to the age at operation and to gender. The secondary aim was to identify the limits regarding the possibility of relocating incongruent joints, both with respect to joint type and age at operation.

Study IV

The aim of this study was to evaluate the long-term results of surgical correction of the shoulder rotational deformity in patients with OBPP, using open subscapularis elongation with or without latissimus transfer. The primary aim was to evaluate whether the improvements in joint mobility and the Mallet score seen at one year post surgery would change over time. The secondary aim was to establish whether the relocated joints would remain stable over time, and whether additional surgery would be necessary after the first year.

PATIENTS & METHODS

ETHICAL CONSIDERATIONS

All studies were conducted according to the principles of the Helsinki Declaration. Ethical approval was obtained from the Regional Ethical Review Board at Karolinska Institutet, 214/99, the Regional Ethical Review Board Gothenburg, S-166-01 and the Regional Ethical Review Board Stockholm, 2011/790-31/4

PARTICIPANTS

The Department of Hand Surgery in Stockholm has served as a national referral centre for obstetric brachial plexus injuries since 1986. By the mid-1990s there was a dammed up need for correction of shoulder rotational deformities, with a large group of patients of varying ages in need of surgery. The participants in the four studies in this thesis were from a series of 276 patients operated on from 1997 to 2009 for correction of a rotational deformity in the shoulder following birth palsy of the plexus.

Studies I and II were conducted on subscapularis muscle biopsy specimens from a subgroup of 13 patients with rotational contractures, who gave their informed consent to participate in the collection of biopsy samples at surgery. Two patients in this group were operated on with a partial tenotomy of the subscapularis and resection of the coracoid only and they were not included in studies III and IV.

For practical reasons, to facilitate the collection and handling of the muscle biopsy samples, they were harvested in three separate series of consecutive operations. Biopsy samples from the 9 first patients were analysed in study I, while all 13 patients were included in study II.

Study I also included biopsy specimens from seven unrelated patients who were unaffected by neuromuscular disorders and who contributed 'control biopsy specimens' from six different uninjured upper limb muscles.

Study III included the 270 patients who were examined at one year after surgery. Four patients were lost to follow-up. The mean age at surgery was 6.2 years (range, 0.6–35) and there were 144 females and 130 males.

Study IV was conducted on a subgroup of surgery patients in whom more than seven years had passed since the original operation. Two-hundred and seven patients were eligible and were contacted with a written request to take part in a follow-up study. One-hundred and eighteen patients were included. Fifteen patients responded but could not come, while 74 patients did not respond. The mean follow-up time was 10.4 years (range 7.0–15.1) and the mean age at examination was 15.1 years (range: 7.6–33.7). There were 64 females and 54 males. In 63 of the 118 examined patients the shoulder joint had been relocated.

SURGICAL CONSIDERATIONS AND TECHNIQUES

Contracture release

Prior to 1997 we had used the subscapularis slide technique[31] to release the rotational contracture. Although the results were good, we had noted that the contracture tended to recur in a high proportion of the patients operated on, an observation that is shared with others.[35,36,46] We were also concerned that the procedure did not allow us to release the contracture in patients who had incongruent joints. Therefore, we adopted the anterior elongation technique described by Birch et al.[28,37] and began to systematically relocate the incongruent joints. With this technique, the approach was via the deltopectoral groove. The tip of the coracoid was carefully excised and the conjoined tendon was split longitudinally but not detached. The subscapularis tendon was divided in a step-cut fashion and then carefully repaired with sufficient elongation to allow 45° of passive external rotation. The capsule had to be opened in all cases with an incongruent joint in order to release the contracture and relocate the head of the humerus, but it was not necessary to incise the joint capsule in the majority of patients with congruent joints.

Latissimus dorsi to infraspinatus transfer

This was performed by routing the detached latissimus dorsi tendon under the posterior part of the deltoid and suturing it to the infraspinatus tendon near its insertion. The patients who were selected for a combined contracture release and transfer either had an active external rotation of less than half of the available passive range, or a power of external rotation of less than MRC grade M4.[68] Testing the power of the external rotation is quite difficult in patients with a rotation contracture because the available passive ROM is very limited. Early in the series we stopped combining joint relocation with a latissimus dorsi transfer because of the strong tendency for the relocated joints to develop a restriction in internal rotation.

The choice of operating procedures, inclusion and exclusion criteria are outlined in Figure 7.

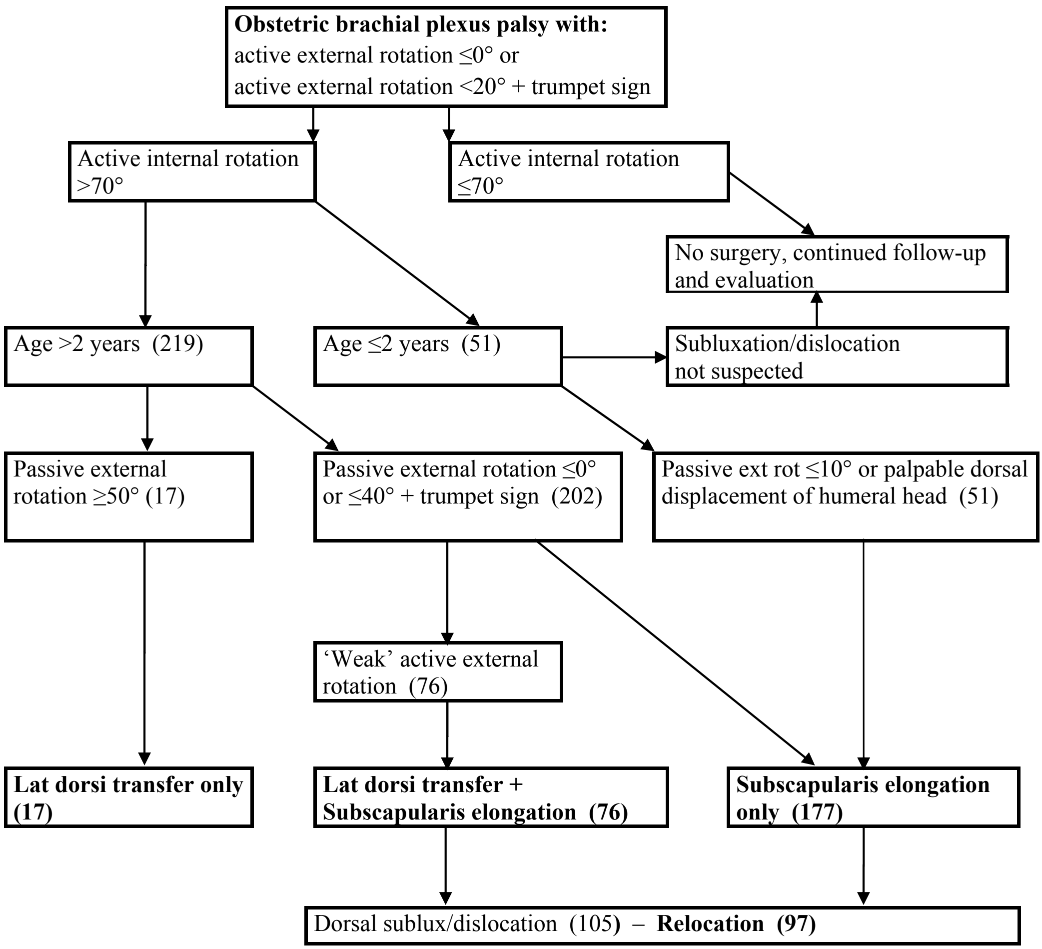

Fig. 7. Treatment algorithm used in study III

A custom-made and individually fitted plastic & aluminium brace with hook-and-loop straps was used to hold the shoulder in adduction and external rotation after surgery (Fig. 8). The families were instructed and trained in the correct application of the brace and were encouraged to remove it daily for purposes of hygiene and changing clothes. The brace was worn for five to six weeks, or seven to eight weeks if the joint had been relocated. Active motion was then resumed and unrestricted activity was allowed three months postoperatively.

Fig. 8. The postoperative brace.

Joint types and relocation

Computerised tomography (CT and later MRI) was used preoperatively in the majority of patients, but our classification was based on *visual inspection of the joint and on its response to the reposition manoeuvre during surgery*. This is an important distinction because even though a radiological examination can give a very good visual representation of a joint deformity it does not necessarily tells us how the joint will behave during a repositioning manoeuvre.

In a *type 1 subluxation* the subluxed head of the humerus rests in the posterior aspect of a deformed glenoid which is flattened or slightly convex. When the arm is rotated externally, the humeral head will slide in a smooth translational movement towards the anterior (anatomical) part of the glenoid (Fig. 9a).

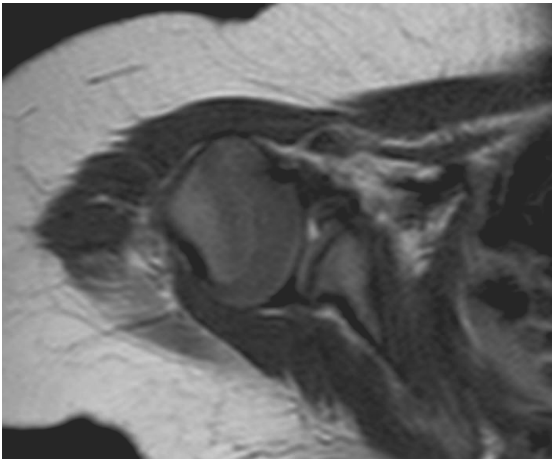

Fig. 9a. MRI image of the glenohumeral joint with a type 1 subluxation.

In a *type 2 subluxation* the glenoid is bifaceted and the subluxed head of the humerus rests in the posterior facet. If the arm is rotated externally, the humeral head will slip over the ridge and click into place in the anterior (anatomical) part of the glenoid. There are two very distinct positions for the humeral head, with a degree of stability in either position (Fig. 9b).

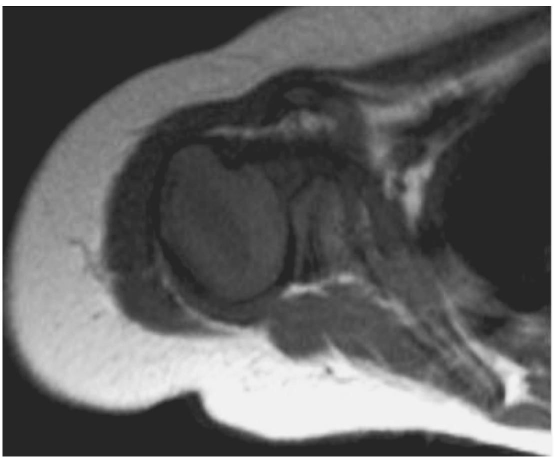

Fig. 9b. MRI image of the glenohumeral joint with a type 2 subluxation.

In a *dislocated joint* the head of the humerus has slipped over the rear edge of the glenoid and rests against the posterior aspect of the scapula. The reposition manoeuvre is similar to that of a bifaceted joint but the humeral head needs to travel further to click into place in the anatomical position. (Fig. 9c).

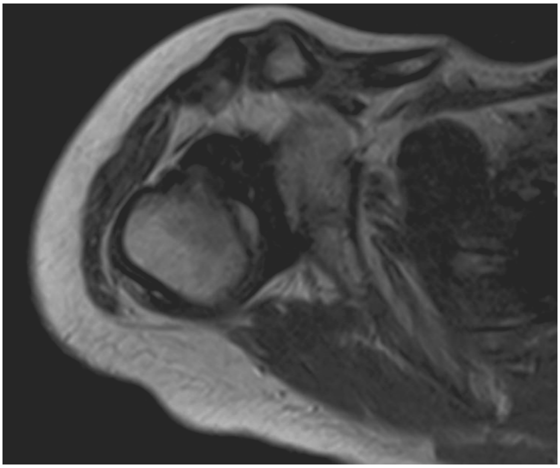

Fig. **9c.** MRI image from a patient with a dislocation.

Rotational osteotomy of the humerus

Historically, this technique was introduced to improve external rotation in situations where interventions on the soft tissues were not deemed possible, e.g. in the presence of a joint deformity. With the introduction of systematic joint relocation the situation has been reversed. The relocation manoeuvre is an external rotation of the arm and when the relocated joints have stabilised a large proportion of them will have. Kambhampati et al.[28] related this phenomenon to retroversion of the humeral head and included a internally rotating osteotomy with the relocation procedure if the estimated retroversion was more than 40°. It was difficult in our series to determine peroperatively if a humeral head was retroverted and therefore elected to do an osteotomy as a second procedure after a minimum of one year. After that time it could be established with better certainty if an osteotomy was required. Thus 23 of the 97 patients with relocated joints in study III were later reoperated on with an internally rotating osteotomy of the humerus. The indication for an osteotomy was an active internal rotation of less than 30°, causing impairment in daily activities. The osteotomy was placed just above the insertion of the deltoid and stabilised with a compression plate. The distal humerus was rotated 40–60°, aiming for a maximum of internal rotation while maintaining a minimum of 15° of external rotation.

Careful measurements before and during the procedure are necessary. A postoperative brace of the same type as in Figure 9 was used for four weeks but with the arm in 0° of rotation.

STUDY DESIGNS

Study I

Previous studies with analyses of the mechanical properties of muscles fibres have shown that patients with contractures due to spasticity (cerebral palsy) have muscle fibres with a shorter resting sarcomere length (SL) and a much higher passive elastic modulus than those of normal controls.[69,70] We wanted to know if similar or other specific changes could be identified in fibres of the subscapularis muscle in patients with OBPP and a shoulder rotation deformity. Muscle fibres from nine patients with OBPP and rotational contracture were studied and compared with biopsy specimens from the upper limbs of seven healthy controls.

Slack SL was determined by transilluminating single muscle fibres or fibre bundles with He-Ne laser beam while floating free in an experimental chamber. This produces a diffraction pattern that can be used to determine the sarcomere lengths.

Muscle fibre specimens were then passively stretched between a force transducer and a micromanipulator and lengthened in 250-µm increments in velocity-independent steps. A tangent modulus for the length/tension curve was calculated for each specimen, using the formula described by Fridén et al.[69] By testing both single fibres and fibre bundles, it was possible to determine changes in both intracellular and extracellular muscle structural components. A total of 31 single muscle fibre and eight fibre bundle experiments were performed on biopsy specimens from the nine patients with OBPP. A total of 20 single muscle-fibre and 12 fibre bundle experiments were performed on the specimens from the seven controls.

Study II

Twenty-four biopsy samples from 13 patients were analysed by two unbiased observers using light microscopy. Specimens were harvested from both the upper and the lower parts of muscles in order to identify possible changes owing to the different innervations of these two areas.

Morphological analyses were done using a light microscope on haematoxylin-eosin-stained sections. The sections were analysed morphologically with respect to the occurrence of central nuclei, angular fibres and necrotic fibres. Fibre diameters were marked by the observers along with fibre circumferences for calculation of sizes and areas using a computerised analysis system. Mean values were calculated from three to four randomly selected areas from each sample. The percentage of extracellular matrix as an indicator of fibrosis was calculated by subtracting the sum of the fibre areas from the total area in a viewing field. The distribution of fast and slow fibres was calculated on sections stained for MHC isoforms. The values obtained were controlled against established references to determine if there were any deviations from the normal.[71]

Study III

This was a case series in which patients with OBPP were studied prospectively, using a pre-determined examination protocol before and one year after surgery for correction of shoulder rotational deformities. We wanted to determine the effect of contracture release using subscapularis elongation and to what extent a balancing muscle transfer (latissimus dorsi to infraspinatus) would contribute to the result. By studying a sufficiently large group of patients we also aimed to determine the effects of interactions due to the condition of the joint (congruent, subluxed, dislocated), age at operation and gender.

One surgeon performed all of the operations. The selection of patients for a combined contracture release and muscle transfer or for contracture release alone was based on an evaluation of the available power of the external rotators, within the available passive rotational range. Patients who had an active external rotation of less than half of the available passive range or who were judged to have a notable weakness in external rotation were selected for the combined procedure. Patients with a lack of active external rotation but no contracture were selected for muscle transfer alone.

The following classifications were used:

Age at operation

1. Under 2 years - The nerve injury has not fully healed
2. 2 to 4 years - Pre-school age
3. 5 to 11 years - Pre-adolescent
4. 12 to 19 years - Adolescent to adult
5. 20 years and over - Adult

Condition of the joint:

1. Congruent
2. Subluxed, type 1 (flattened or rounded glenoid)
3. Subluxed type 2 (bifaceted glenoid)
4. Dislocated

Type of surgery:

1. Subscapularis elongation only
2. Subscapularis elongation + latissimus dorsi transfer
3. Latissimus dorsi transfer only

In the statistical analysis 'joint relocation' was not categorised as a surgical procedure. Since relocation was performed or attempted in all incongruent joints we chose to only categorise the joints according to their type. Also we did not differentiate between successful and failed relocation in the mixed models analysis. Rather than adding a new level and ending up with eight joint categories in the statistical calculations, we elected to let the eight patients in whom relocation failed remain in their original categories for the statistical analysis. A separate calculation of mean values and CIs was presented for the group with failed relocations.

Thus two surgical techniques with three possible combinations, four types of joints and five age groups were the categories used in the mixed effects models analysis in study III.

The numbers of patients in each category are presented in tables 1 and 2 in study III.

Outcome measures

Active ROM for shoulder rotation, abduction and flexion was recorded with a goniometer and with pre-determined reference and sighting lines. All measurements were recorded as the thoraco-humeral mobility

The Mallet score was recorded using the 3-grade scale, where each of the five specified movements is graded from 1 to 3, giving a total score of from 5 to 15.[46]§ Hand-to-mouth ('trumpet sign') and hand-to-head movements were also recorded as separate movements with scores of 1 to 3 respectively.

Because of the extended time period (13 years) needed to collect data from a sufficient number of patients, it was impractical to have an independent person perform all of the pre- and postoperative measurements of ROM, so these were done by the surgeon (TH). Pre- and post-operative Mallet scores were recorded independently by occupational therapists.

Study IV

This was a study of the long-term results of surgery in a subgroup of patients from study III who had a follow-up time of seven years or more after the initial operation.

The following classifications were used:

As there were fewer patients in the long-term study, the number of subgroups was reduced to obtain a sufficient number of subjects in each subgroup. The short-term study had not shown any clinically relevant differences in the results for subjects who were operated on between the ages of 2 and 19 and therefore these patients were combined into one category. Thus the age groups in study IV were reduced to three:

Age at operation

1. Under 2 years
2. 2 to 19 years
3. 20 years and over

Twenty-three of the patients who had had their joints relocated were reoperated on after the first year with an internally rotating osteotomy of the humerus. Three new joint categories were created according to whether or not an osteotomy had been performed:

1. Congruent
2. Relocated
3. Relocated with subsequent rotational osteotomy

Study IV included eight patients who originally had been operated on with a latissimus dorsi transfer alone. Five of them had developed a rotational contracture after the first year and had been reoperated on with a subscapularis elongation. All of the eight patients included remain in their original subgroup (‘transfer only’) in the statistical analysis. The numbers of patients in each category are presented in tables 1 and 2 in study IV.

Outcome measures

The same outcome measures as in study III were used. The examinations were conducted over a two-year period and all measurements were recorded by three surgeons who had not been involved in the surgery. A questionnaire was used to assess whether there were functional impairments due to a limitation of internal rotation in the shoulder. The participants were requested to grade their ability to perform three activities requiring internal rotation. Each activity was given three grades: easy, difficult or impossible.

The results of these questionnaires were displayed in bar charts which also included the corresponding values for internal rotation in order to visualise whether there was a correlation between the range of internal rotation and impairment of the specified function.

STATISTICAL METHODS

Study I

The tangent modulus for the length/tension curve of the fibres and fibre bundles was calculated using the equation for fibre elasticity published by Fridén et al.[69] The data were grouped by experiment type (single fibre versus fibre bundle) and sample type (control versus OBPP) and analysed using a two-sided independent samples T-test. The data were presented as the mean and standard error of the mean (SEM).

All analyses were performed using Statview version 5.0 (SAS Inst. Inc. Cary, NC, USA).

Study II

Analyses and calculations were performed using a computerised analysis system (Easy Image measure module 2000, Bergström Instrument AB, Stockholm, Sweden).

The morphological analyses were performed by two independent observers. The inter-observer error was calculated as the deviation in percent in the values for the less experienced observer from those of the more experienced observer for each sample. The mean and standard deviation for the absolute values of these differences were then calculated as a measure of agreement.

A linear regression analysis was used to investigate the relationship between fibre size and age. Significance was set at $p < 0.05$. All analyses were performed using Statview version 5.0 (SAS Inst. Inc. Cary, NC, USA).

Study III

A mixed effects models regression analysis was used to evaluate the effects of surgery on ROM and the Mallet score, because it is well suited for the analysis of repeatedly measured outcomes when there are missing data and when the repeated measures are irregularly spaced over time and the sample sizes are modest to large.[72]

Independent factors were operative status (pre- and post- surgery), gender, age, condition of the joint and the type of surgical intervention (elongation only, elongation plus transfer or transfer only).

Eight patients in whom relocation of the joints failed, were included in the calculations but they were also studied as a separate group, with the pre- to post-operative changes calculated using the paired samples t-test. All tests were two-sided and the results were considered significant at $p < 0.05$. All analyses were performed using IBM SPSS version 20.0 (IBM Corp., Armonk, New York).

Study IV

A mixed effects models regression analysis was used to evaluate the effects of surgery on ROM and the Mallet score. Independent factors were operative status (pre-, 1 and >7 years post- surgery), gender, age, joint type (congruent, relocated, relocated plus osteotomy) and transfer or no transfer.

All tests were two-sided and the results were considered significant at $p < 0.05$. All analyses were performed using IBM SPSS version 21.0 (IBM Corp., Armonk, New York).

The results of the mixed models analysis in both study III and study IV were displayed in tables. Raw means and 95% CIs for ROMs and Mallet scores were also presented in bar charts to better visualise the changes from pre- to post-surgery.

The pre-operative and one-year post-operative values presented all refer to the larger group of patients (n = 270) examined at those time points.

RESULTS

STUDY I

The slack SL in single fibres from OBPP patients, 2.01 µm (SEM, 0.03 µm), was significantly ($p < 0.01$) shorter than in normal controls: 2.26 µm (0.08 µm). This difference could not be seen between fibre bundles. The range of SL with linear deformation of the fibres was significantly ($p < 0.01$) wider for OBPP patients: 2.09 µm (0.09 µm) than for normal controls: 1.75 µm (0.07 µm). No differences were seen between fibre bundles.

The tangent modulus for both single fibres and fibre bundles showed higher means for OBPP patients compared with normal controls, but the difference was not significant. The relative increase in stiffness between single fibres and fibre bundles was 6.7 times for OBPP samples and 5.3 times for controls. This constitutes a 26% larger increase in stiffness for the OBPP samples compared with normal controls. The tangent modulus for fibre bundles corrected for the number of fibres in the bundle was calculated without any significant difference between OBPP patients and controls, viz. 8.0 (3.1) versus 6.7 (2.3) kPa.

The changes are consistent with the theory that secondary changes in muscle fibre properties occur as a result of a lack of sufficient passive stretch and that there may be compensatory changes in the extracellular matrix. The results suggest the presence of a dynamic feedback system constituting a muscle-to-extracellular matrix communication interface.

STUDY II

The muscle morphology was considered normal in 12 of the 13 patients. The mean fibre occupation for the 12 patients was 93 (0.4) %. The mean fractions of angular and necrotic fibres were 1.3 (0.5) % and 1.0 (0.3) %, respectively. The mean value for fibres with central nuclei was 9.3 (3.9) %. Five patients had central nuclei levels of less than 3%, four had levels between 3% and 10% and three patients had levels of more than 10%. There was a positive regression ($r^2 = 0.76$) between age and fibre diameter, indicating a significant ($p < 0.001$) linear relationship. These patients had a predominance of MHC type I (slow) fibres with a mean of 75 (3.8)%. Fibre type grouping (regarded as a sign of reinnervation) was not detected in any of the 12 biopsy samples.

One of the patients displayed very different muscle morphology, with an abnormal mean fibre diameter relative to age and a mean fibre occupation of 79%, indicating lack of growth and excessive fibrous tissue. No difference in the occurrence of angular and necrotic fibres was noted in the samples from this subject compared with the other 12 patients. Central nuclei were found in 4.7% of the fibres. The patient in question had a type II (fast) fibre proportion of 66% and there were signs of regrouping of fibres.

The results support the theory that the contracture develops as a result of muscular imbalance in the majority of patients, although the findings in one of the biopsy samples suggest that fibrosis formation can also be a factor.

STUDY III

Primary outcomes:

1. Subscapularis elongation provided an overall increase in the mean external rotation of 84.6° (95% CI: 80.2 to 89.1, $p < 0.001$) at the one-year follow-up. There was a substantial improvement in all age groups under 20 years (Fig. 10).

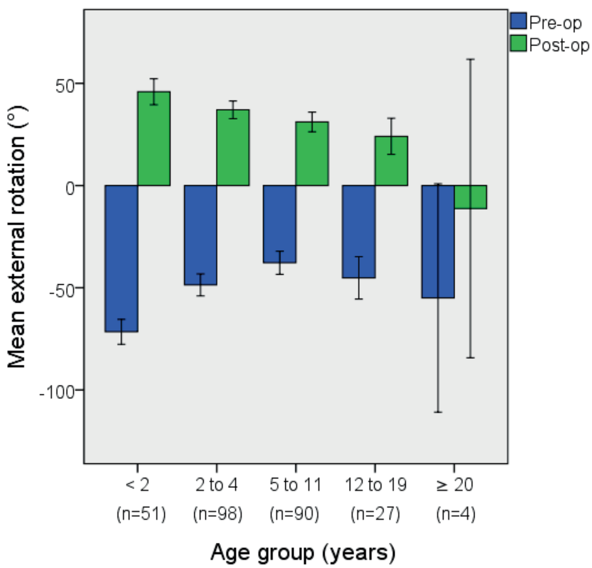

Fig. 10. Bar charts showing mean pre- and post-operative active external rotation by age group. The error bars denote the 95% confidence intervals.

2. There was no significant difference in the improvement in mean external rotation between the combined subscapularis elongation plus latissimus dorsi-to-infraspinatus transfer group and the elongation only group.
3. When the latissimus dorsi was transferred alone, in patients without contracture, the mean external rotation at one year post surgery was 9.4° (95% CI: -27.3 to 8.5, $p = 0.3$) less than in the elongation only group.
4. If the shoulder joint was congruent, there was a mean adjusted loss of internal rotation of 8.6° (95% CI: -12.0 to -5.2, $p < 0.001$)
5. In the incongruent joint groups, the adjusted mean loss of internal rotation was between 28° (95% CI: -36.6 to -18.5, $p < 0.001$) and 34° (95% CI: -40.4 to -29.0, $p < 0.001$), depending on the condition of the joint. (Fig.11).

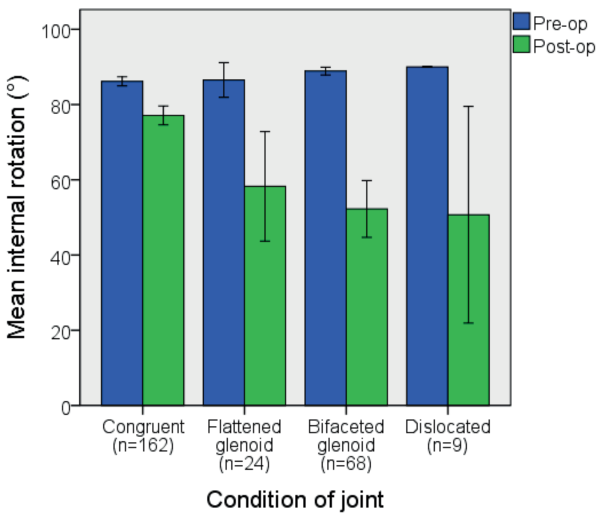

Fig. 11. Bar charts showing mean pre- and post-operative active abduction by joint category. The error bars denote the 95% confidence intervals.

6. There were no significant changes in shoulder abduction or flexion from pre- to post-surgery in any group.
7. The mean Mallet aggregate score improved by 4.0 points (95% CI: 3.7 to 4.2, $p < 0.001$).

8. The trumpet sign was corrected (from Mallet grade 1 or 2 to Mallet grade 3) in 156 of 188 patients (83%).

Secondary outcomes:

1. Subluxed shoulder joints could be relocated up to the age 12 years and dislocated joints up to the age of 5 years.

STUDY IV

Primary outcomes:

1. The overall mean improvement in external rotation from pre-surgery to the long-term follow-up was 66.5° (95% CI: 61.5 to 71.6, $p < 0.001$).
   External rotation by joint category is presented in Figure 12.

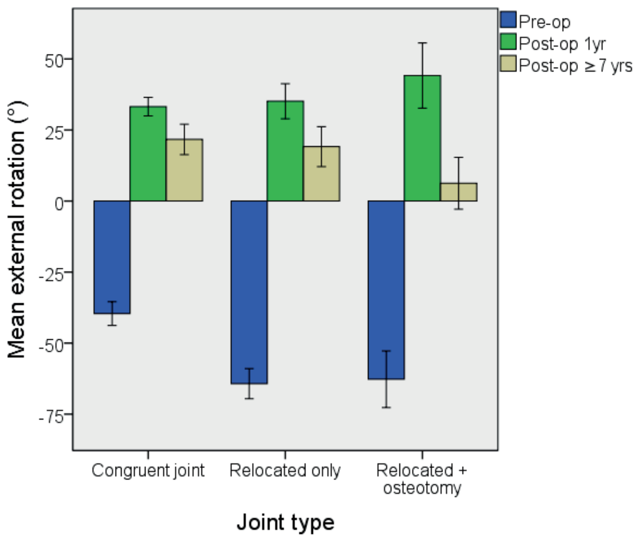

Fig. 12. Bar chart showing the mean external rotation by joint category at the three time points. The error bars denote the 95% confidence intervals.

2. There were no statistically significant differences in mean external rotation between the elongation only, elongation plus transfer and the transfer only groups at the long-term follow-up.
3. The overall mean internal rotation had decreased by a mean of 22.6° (95% CI: -18.7 to -26.5, $p < 0.001$). There were no statistically significant differences in active internal rotation between the three joint types at the long-term follow-up. (Fig. 13).

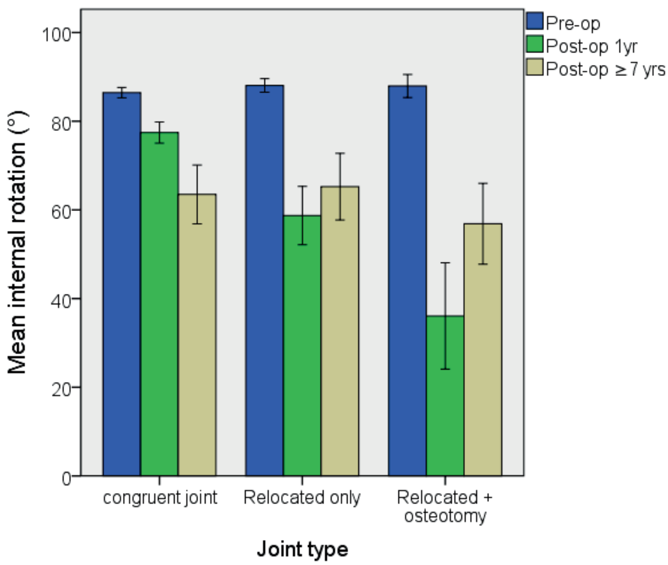

Fig. 13. Bar chart showing the mean internal rotation by joint category at the three time points. The error bars denote the 95% confidence intervals.

4. There was an overall loss of mean abduction from pre-surgery to the long-term follow-up of 7.4° (95% CI: -2.0° to -12.7°; $p = 0.007$) at the long-term follow-up.
5. The mean improvement in the 3-grade Mallet score from pre-surgery to the long-term follow-up was 3.1 units (95% CI 2.7 to 3.4, $p < 0.001$), from 8.7 (95% CI: 8.4 to 9.0) to 11.8 (11.5 to 12.1).
6. At the long-term follow-up, the trumpet sign was corrected (from Mallet grade 1 or 2 to Mallet grade 3) in 71 of 90 patients (79%).

Secondary outcomes:

1. Twenty-three of the 97 patients with relocated joints had been reoperated on with an internally rotating osteotomy to correct an excessive loss of internal rotation. In five of them there had been a recurrence of (intermittent) subluxation, at between three and six years after the first operation and in three of these patients the reoperation also included a stabilisation procedure on the posterior joint capsule. All joints were stable at the long-term follow-up.
2. Five of eight examined patients who originally had been operated on with a latissimus dorsi transfer alone had subsequently developed a contracture which had necessitated reoperation with subscapularis elongation.
3. The results of the questionnaires suggested that the loss of internal rotation in this group of patients did not present a major problem when buttoning and unbuttoning a shirt or a blouse but a group of patients with very limited internal rotation could not reach their opposite armpit without the aid of the unaffected hand.

CONCLUSIONS

Biopsy specimens from the subscapularis muscle in patients with a rotational contracture in OBPP display changes that are consistent with the theory that the deformity is the result of a neuromuscular imbalance rather than direct trauma to the muscle in the majority of patients.

Open subscapularis elongation effectively releases the rotational contracture and corrects the trumpet sign in patients with OBPP, up to the age of 20 years, with an acceptable mean loss of internal rotation. There is a moderate reduction in the rotation sector over time following surgery.

Adding a latissimus dorsi to infraspinatus transfer does not improve the external rotation in patients in whom the subscapularis tendon is elongated due to contracture. A latissimus dorsi-to-infraspinatus transfer can provide useful external rotation in patients with a lack of muscle power and without contracture but it cannot be relied upon to prevent future development of the contracture.

Open reduction of a dorsally subluxed or dislocated joint is generally possible in patients up to the age of 12 and 5, respectively. Up to a third of these patients can be expected to require additional surgery in order to adjust the rotation sector and/or to stabilise the shoulder joint.

There are very large individual variations and postoperative monitoring should continue for many years because of the need for additional surgery in selected patients.

GENERAL DISCUSSION

The rotational deformity of the shoulder in obstetric palsy is a complex mix of muscular weakness and muscle shortening. It is often further complicated by skeletal deformities in the shoulder area and incongruence of the glenohumeral joint. The wide variation in the severity of the nerve injury and in the degree of impact on muscle function and joint development makes it very difficult to establish simple guidelines for treatment. The low incidence of the brachial plexus birth injury is fortunate but it also contributes to making it a difficult problem to study.

WHAT IS THE MECHANISM BEHIND THE DEFORMITY

We tend to think of the obstetric plexus lesions as primarily a problem in the nerves. Yet the nerve injuries lead to profound changes in all of the affected tissues: in muscles, tendons, joint capsules, joints and bones. Muscles are not mere slaves to the input from the nervous system; the muscles themselves have individual characteristics which determine their response to nerve signals. Following a severe nerve injury there will be secondary changes in the muscles themselves which alter their response to nerve signals.

The shoulder deformities that follow OBP lesions usually have a characteristic pattern with weakness in external rotation and contracture of the subscapularis but there are large individual variations. Fairbank[22] and Sever[10] noted that many children with the rotation deformity not only had the dominant contracture of the subscapularis but also had a degree of stiffness in other muscles of the rotator cuff. This is difficult to explain if we subscribe to the theory that the contractures are the result of muscular imbalance alone.

Zancolli identified two groups of OBPP patients with shoulder sequelae.[45] A larger group included children with a shoulder contracture, which he believed was the result of an Erb's palsy *combined with* an obstetric trauma of the shoulder joint and other surrounding soft tissues. In a smaller group of approximately 10% of the OBPP patients, Zancolli observed a flaccid paralysis without contracture and he believed that in these children there was only a nerve trauma, without other associated soft tissue injuries.

The two biopsy studies in this thesis were aimed at identifying mechanical or morphological changes in the contracted subscapularis muscles of OBPP patients that might support either of the above theories.

Study I verified that there were mechanical changes in the subscapularis biopsy specimens from OBPP children, which separated them from normal controls and from the biopsies of spastic muscles which had been studied previously.[69,70] Compared to those of the spastic muscles, however, the mechanical changes in the OBPP biopsies were relatively discrete and they do not support the theory that the contracture develops as a result of fibrosis.

The morphological changes that we observed in study II were also very mild. The majority of the biopsy specimens had an essentially normal morphology and we could not detect signs of increased fibrosis except in one. The results are consistent with the theory that the rotational contracture is an effect of muscular imbalance in the majority of cases but they are not conclusive.

Recent studies[58,59] have shown that experimentally incurred plexus injuries in mice caused muscle shortening and contractures in the shoulder and elbow and the explanation for this would be an impairment of muscle growth. The contractures were inversely correlated with the degree of the nerve regeneration. Thus the worst contractures were seen in mice with very little recovery of muscle function. The theory of impaired muscle growth based on experiments on mice adds a new dimension to the problem but there are also reasons to question this theory. It does not explain why the loss of growth would consistently be most severe in the subscapularis or why we see so many children with excellent neurological recovery but with a severe contracture of the subscapularis. The worst type of brachial plexus lesion, with avulsion of the spinal nerves produces a flaccid paralysis without contractures.

Impaired muscle balance and muscle control most probably play a part in the formation of the contracture. It is not a simple question of balance: it is a matter of imbalance between muscles that are compromised by a nerve injury. Study IV in this thesis demonstrated that a latissimus dorsi-to-infraspinatus transfer in OBPP patients with weak external rotation, but no contracture, could prevent future development of a contracture in only half of the patients so operated on. We are trying to help a dysfunctional muscle by adding a second muscle that may also be dysfunctional.

Most likely, there are other mechanisms involved too and there may even be different mechanisms at work at different stages of development and different ages.

One phenomenon in OBPP, which has not been studied well and which may contribute to contracture formations over time, is the lack of co-ordination in the affected arm. OBPP patients usually cannot swing the affected arm in a normal way when they run, but tend to lock it against the trunk. Frequently, when these patients are requested to do a specific movement in the examination room, they activate antagonistic muscles which cause the limb to tense up and to limit the mobility. Many children with OBPP appear to have a lack of co-ordination with unnecessary and unwanted activity in muscles that should normally be relaxed in well-controlled motions. This limits the useful ROM and may conceivably contribute to contracture formation over time.

The phenomenon of involuntary activation of antagonistic muscles is common in OBPP and is termed 'co-contraction.' The mechanisms behind this are not well understood and merit further studies.

THE RESULTS OF SURGERY

The results of the surgical treatment presented in studies III and IV compare favourably with those reported in other studies. The increase in the Mallet score and in external rotation in our studies was similar to those observed in two other studies using the same technique. In their series of 183 relocated joints, Kambhampati et al.[28] reported increases in the mean Mallet score from 9.4 to 13 and in mean external rotation of 58°. In their smaller series of 19 patients, van der Sluijs et al.[38] reported an increase in mean passive external rotation of from -9.7° to 58.4°.

A recent study by Dodwell et al.,[43] which looked at the results at a mean of 20 months after combined subscapularis slide, latissimus dorsi and teres major transfers and glenoid anteversion osteotomy, presented results which are very similar to ours. There was a gain in mean active external rotation of 82°, a loss of mean active internal rotation of 26° and an improvement in the mean Mallet aggregate (5 grade) score of 4.0 points.

The reported loss of internal rotation varies a great deal in published studies. In their series of 183 relocated joints, 70 of which had had been combined with an internally rotating humeral osteotomy, Kambhampati et al.[28] reported a mean loss of only 10°. In their series of 19 patients with mixed joint types, van der Sluijs et al.[38] reported that 42% of them needed an osteotomy but did not report the actual loss of active internal rotation. Pearl et al.[32], in a series of 19 children who underwent arthroscopic release, reported a mean loss of 37° of internal rotation after release only and a loss of 42° in patients with a combined release and latissimus dorsi transfer. In a long-term study by Kirkos et al.,[73] ten patients who had been operated on with a subscapularis tenotomy and latissimus dorsi transfer were followed for a mean of 30 years. There was a preoperative mean internal rotation of 44°, which was only slightly reduced to 39° at 30 years post surgery.

Studies that report the postoperative improvement in passive motion only are difficult to interpret.[38] Passive motion is indeed a measure of how well a contracture has been released but it does not describe how well the limb performs. Patients with OBPP often cannot make full use of their passive movement range, so the active ROM needs to be evaluated too.

The Mallet test for internal rotation appears to be a little too insensitive. In our series we saw no change in the mean Mallet score for hand-to-back, even though there was an overall reduction in the mean active internal rotation of 22.6° from pre-surgery to the long-term follow-up.

THE TRUMPET SIGN

Correcting the trumpet sign is a major factor in improving limb function in OBPP patients with the rotation contracture (Fig. 14).

If there is one single measure that might sum up the efficiency of a treatment for the rotation contracture, correction of the trumpet sign would be a good candidate. It is a dominant feature of the contracture and because it is so visible, it is not only a functional problem but can also be disturbing in a social context.

Presenting the result of surgery as the percentage of patients operated on who achieve full correction of the trumpet sign, i.e. from 1 or 2 to 3 in the 3-grade score, may be more relevant than the change in the mean value.

Fig. 14. A marked trumpet sign in an adolescent girl. Complete correction one year after subscapularis elongation.

STRENGTHS AND LIMITATIONS

Studies I and II

The strength of these studies is that they were done on biopsy specimens from affected patients. There are few such studies. A major limitation is the lack of adequate reference biopsies, which makes it difficult to draw conclusions. It would be possible to collect samples from other muscles in OBPP patients subjected to surgery, but obtaining reference samples from unaffected shoulder muscles presents a problem. Furthermore it is difficult to determine the causality in changes that can be observed in isolated biopsy samples. We cannot say if the fibrosis that was noted in one patient was the result of the birth injury or a long-standing dislocation.

Studies III and IV

The strength of these two studies is that the patients were studied prospectively, with a defined treatment algorithm and using a limited set of surgical procedures, all of which were performed by one surgeon. Pre- and post-operative ROM measurements were done using the same protocol with defined reference points and reference lines.

Both studies included large series of patients in comparison with many other published studies and the follow-up time in study IV was also comparatively long.

The Mallet scores in both studies, as well as ROM measurements in study IV, were recorded by persons who had not been involved in the surgery.

An important limitation of study III and study IV is the lack of control groups. They are both uncontrolled before and after studies. Part of the improvement from pre- to one year post-surgery in the youngest age group is most likely a result of their increasing maturity and an improved neuromuscular function owing to continuous nerve healing.

Patients with contractures were not randomised into the two groups of transfer vs. no transfer; they were actively selected to either group by the surgeon, based on our conviction at the time that many of these patients needed a transfer. Thus, the evaluation of the results with respect to transfer or no transfer becomes difficult. In hindsight, it can be said that despite our efforts to select patients, the results show remarkably small differences in ROM between the two groups, both pre- and post-operatively. The most likely explanation is that the addition of a muscle transfer contributed very little to the result of the contracture release.

A limitation of study III is that pre- and postoperative examinations were conducted by the surgeon, which introduces a risk of bias and for systematic error. The latter is outweighed to some extent by a strict examination protocol.

A limitation of study IV is that a large proportion of the patients were lost to the follow-up. There was an inclusion bias with a larger proportion of patients with relocated joints in the examined group than in the group that was lost to follow-up. An analysis of the respective results for the two groups at the short-term follow-up showed lower mean values in abduction and internal rotation for the group that was later examined in study IV. That suggests that the results of the long-term follow-up were not exaggerated.

IMPLICATIONS FOR CLINICAL PRACTICE

Open subscapularis elongation provides an efficient release of the shoulder rotation contracture in OBPP and offers the opportunity to relocate incongruent joints within certain given age limits.

It does not appear possible to determine the function of the external rotators in the presence of an internal rotation contracture. Adding a muscle transfers does not appear to improve the results when a contracture is released and the combined procedure has therefore been abandoned.

Patients who have a lack of external rotation but no contracture benefit from a latissimus dorsi transfer but approximately half of them can be expected to develop a contracture eventually. Therefore these patients need to be monitored for many years following such a procedure.

One third of the patients in whom the glenohumeral joint is relocated can be expected to recover less than 30° of internal rotation and need to be considered for an internally rotating osteotomy after the first postoperative year. A smaller group of patients can be expected to develop a recurrent subluxation several years after relocation. Patients with relocated joint therefore need to be monitored for many years postoperatively.

Even though the loss of mean internal rotation after subscapularis elongation appears to be only moderate, there is a large individual variation and the procedure is not recommended for patients who already have a limited internal rotation, who have poor shoulder flexion or a lack of wrist flexion. Patients should be informed that there may be a loss of internal rotation that requires additional surgery.

The aggregate Mallet score has limited use in evaluating the rotational deformity of the shoulder in OBPP patients. Careful documentation of active and passive ROM combined with evaluation of the selected movements for hand-to-mouth and hand-to-head seems to provide a good basis for the evaluation.

FUTURE RESEARCH

Further studies on the mechanisms behind the shoulder rotational deformities in OBPP should contribute to a better understanding of the problems at hand. An evaluation the pattern and distribution of the motor endplates in the subscapularis, if compared with biopsies from the infraspinatus muscles, could provide further clues to differences in the reinnervation between external and internal rotators.

It may also be possible to further investigate the phenomenon of co-contractions by using neurophysiological techniques.

It seems unlikely that we will see a radical change in the treatment arsenal in the near future. It would be beneficial to further establish the potential and the limitations of the surgical procedures that are available today. In which situations, for instance, is a subscapularis release plus relocation adequate and when is a posterior wedge osteotomy or glenoplasty indicated?

This thesis has not considered the radiological evaluation of the relocated joints. Such studies are needed in order to determine the long-term effect of joint relocation with respect to joint development and remodelling and in order to identify possible related problems.

Changes in ROM, and in sum scores such as the Mallet test can certainly be used to detect statistically significant effects of surgery but it is important to be clear about which improvements that are relevant for the patients and how well they hold up over time. There is a need for studies that can establish the value of specific surgical interventions for the patients in their daily activities and in the long term.

ABSTRACT IN SWEDISH

En felställning med inåtroterad axelled är mycket vanlig hos patienter med obstetrisk plexus brachialis-skada. Trots att förvånansvärt korrekta beskrivningar av felställningen publicerades redan i början på 1900-talet, så är mekanismerna bakom den inte klarlagda och det finns ingen konsensus angående den kirurgiska behandlingen.

Studierna i denna avhandling syftade till att förbättra det vetenskapliga underlaget för kirurgisk behandling av rotationsfelställningen efter obstetrisk plexusskada.

I studie I undersöktes de passiva mekaniska egenskaperna hos enstaka fibrer och fiberbuntar från muskelbiopsier tagna ur subscapularis hos nio barn som opererades för rotationskontraktur. Biopsier från sju friska individer användes som kontroller. Enskilda muskelfibrer från patienter med obstetrisk plexusskada uppvisade kortare vilolängd i sarcomererna och en linjär deformation av fibrerna inom ett större intervall av sarcomerlängd än kontrollerna. Preparaten från OBPP-patienter uppvisade också en större relativ ökning av styvheten hos fiberbuntar jämfört med enstaka fibrer.

I studie II undersöktes histopatologiska förändringar hos muskelbiopsier som tagits från subscapularismuskeln från 13 barn med plexusskada, i samband med kirurgi. Majoriteten av dessa biopsier hade en i huvudsak normal morfologi och uppvisade en övervikt av MHC typ I, dvs. långsamma fibrer. Resultaten av studie I och II överensstämmer med teorin att förkortningen i subscapularismuskeln hos majoriteten av patienterna är en effekt av ändrad muskelbalans till följd av nervskadan.

I studie III utvärderades 270 patienter med förlossningsskada i plexus och rotations-felställning, ett år efter kirurgisk korrektion med öppen subscapularisförlängning och / eller transferering av latissimus dorsi till infraspinatus. Reposition utfördes av alla inkongruenta leder och var framgångsrik hos 92% (97 av 105) av patienterna, med en övre åldersgräns på 12 år för subluxerade leder och 5 år för luxerade leder. Kirurgin medförde en stor förbättring i genomsnittlig utåtrotation och Mallet score. En påtaglig inskränkning i inåtrotation noterades för patienterna vars leder hade reponerats men inte för dem med kongruenta leder. Trumpettecknet korrigerades hos 83%. Subscapularis-förlängning kombinerad med latissimus dorsi transferering leddes inte till bättre resultat än när enbart förlängningen gjordes.

I studie IV utvärderades långtidsresultaten (7 år eller mer efter kirurgin) hos 118 av patienterna från studie III, med hjälp av samma undersökningsprotokoll. Studien visade en måttlig genomsnittlig minskning av rotationssektorn jämfört med ettårsresultaten, medan den genomsnittliga abduktionen kvarstod oförändrad liksom korrektionen av trumpettecknet. En fjärdedel av patienterna med reponerade leder hade reopererats för att justera rotationssektorn eller stabilisera axelleden.

Slutsatserna av studierna var att öppen subscapularisförlängning gav god långsiktig förbättring av den aktiva utåtrotationen med kvarstående reposition av axellederna och korrektion av trumpettecknet och med en måttlig genomsnittlig inskränkning av inåtrotationen. Långsiktig uppföljning av de opererade patienterna rekommenderas pga. stora individuella variationer och behovet av kompletterande kirurgi hos utvalda patienter.

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REFERENCES

1. Sjoberg I, Erichs K, Bjerre I. Cause and effect of obstetric (neonatal) brachial plexus palsy. *Acta Paediatr Scand 1988;77-3:357-64.*
2. Bager B. Perinatally acquired brachial plexus palsy--a persisting challenge. *Acta Paediatr 1997;86-11:1214-9.*
3. Mollberg M, Hagberg H, Bager B, Lilja H, Ladfors L. High birthweight and shoulder dystocia: the strongest risk factors for obstetrical brachial plexus palsy in a Swedish population-based study. *Acta Obstet Gynecol Scand 2005;84-7:654-9.*
4. Gherman RB, Ouzounian JG, Goodwin TM. Brachial plexus palsy: an in utero injury? *Am J Obstet Gynecol 1999;180-5:1303-7.*
5. Ecker JL, Greenberg JA, Norwitz ER, Nadel AS, Repke JT. Birth weight as a predictor of brachial plexus injury. *Obstet Gynecol 1997;89-5 Pt 1:643-7.*
6. Mollberg M, Hagberg H, Bager B, Lilja H, Ladfors L. Risk factors for obstetric brachial plexus palsy among neonates delivered by vacuum extraction. *Obstet Gynecol 2005;106-5 Pt 1:913-8.*
7. Nath RK, Kumar N, Avila MB, Nath DK, Melcher SE, Eichhorn MG, Somasundaram C. Risk factors at birth for permanent obstetric brachial plexus injury and associated osseous deformities. *ISRN Pediatr 2012;2012:307039.*
8. Weizsaecker K, Deaver JE, Cohen WR. Labour characteristics and neonatal Erb's palsy. *BJOG 2007;114-8:1003-9.*
9. Mollberg M, Lagerkvist AL, Johansson U, Bager B, Johansson A, Hagberg H. Comparison in obstetric management on infants with transient and persistent obstetric brachial plexus palsy. *J Child Neurol 2008;23-12:1424-32.*
10. Sever JW. Obstetric paralysis - its cause and treatment. *Can Med Assoc J 1920;10-2:141-61.*
11. Strombeck C, Krumlinde-Sundholm L, Forssberg H. Functional outcome at 5 years in children with obstetrical brachial plexus palsy with and without microsurgical reconstruction. *Dev Med Child Neurol 2000;42-3:148-57.*
12. Pondaag W, Malessy MJ, van Dijk JG, Thomeer RT. Natural history of obstetric brachial plexus palsy: a systematic review. *Dev Med Child Neurol 2004;46-2:138-44.*
13. Gilbert A, Tassin JL. [Surgical repair of the brachial plexus in obstetric paralysis]. *Chirurgie 1984;110-1:70-5.*
14. Bisinella GL, Birch R. Obstetric brachial plexus lesions: a study of 74 children registered with the British Paediatric Surveillance Unit (March 1998-March 1999). *J Hand Surg [Br] 2003;28-1:40-5.*
15. Hoeksma AF, Wolf H, Oei SL. Obstetrical brachial plexus injuries: incidence, natural course and shoulder contracture. *Clin Rehabil 2000;14-5:523-6.*
16. Waters PM. Comparison of the natural history, the outcome of microsurgical repair, and the outcome of operative reconstruction in brachial plexus birth palsy. *J Bone Joint Surg Am 1999;81-5:649-59.*
17. DiTaranto P, Campagna L, Price AE, Grossman JA. Outcome following nonoperative treatment of brachial plexus birth injuries. *J Child Neurol 2004;19-2:87-90.*
18. Geutjens G, Gilbert A, Helsen K. Obstetric brachial plexus palsy associated with breech delivery. A different pattern of injury. *J Bone Joint Surg Br 1996;78-2:303-6.*
19. Kennedy R. Suture of the brachial plexus in birth paralysis of the upper extremity. *Br M J 1903;1(2197):298-301.*

20. Clark LP, Taylor A.S.,Prout,T.P., . A study on brachial plexus palsy. *Am J Med Sci 1905;130-4:670-707.*
21. Gilbert A, Khouri N, Carlioz H. [Birth palsy of the brachial plexus--surgical exploration and attempted repair in twenty one cases (author's transl)]. *Rev Chir Orthop Reparatrice Appar Mot 1980;66-1:33-42.*
22. Fairbank HAT. Birth Palsy: subluxation of shoulder joint in infants and young children. *Lancet 1913;3-1:1217-23.*
23. L'Episcopo JB. Restoration of muscle balance in the treatment of obstetrical paralysis. *N Y J Med 1939-39:357-63.*
24. Rogers MH. An operation for the correction of the deformity due to "obstetrical paralysis". *Boston Med Surg J 1916-174:163-4.*
25. Whitman R. VIII. The Treatment of Congenital and Acquired Luxations at the Shoulder in Childhood. *Ann Surg 1905;42-1:110-5.*
26. Lin JC, Schwentker-Colizza A, Curtis CG, Clarke HM. Final results of grafting versus neurolysis in obstetrical brachial plexus palsy. *Plast Reconstr Surg 2009;123-3:939-48.*
27. Gilbert A, Pivato G, Kheiralla T. Long-term results of primary repair of brachial plexus lesions in children. *Microsurgery 2006;26-4:334-42.*
28. Kambhampati SB, Birch R, Cobiella C, Chen L. Posterior subluxation and dislocation of the shoulder in obstetric brachial plexus palsy. *J Bone Joint Surg Br 2006;88-2:213-9.*
29. Strombeck C, Krumlinde-Sundholm L, Remahl S, Sejersen T. Long-term follow-up of children with obstetric brachial plexus palsy I: functional aspects. *Dev Med Child Neurol 2007;49-3:198-203.*
30. Hultgren T, Jönsson K, Pettersson H, Hammarberg H. Surgical correction of a rotational deformity of the shoulder in patients with obstetric brachial plexus palsy. Short-term results in 270 patients. *Bone Joint J 2013;95-B:in press.*
31. Carlioz H, Brahimi L. [Place of internal disinsertion of the subscapularis muscle in the treatment of obstetric paralysis of the upper limb in children]. *Ann Chir Infant 1971;12-2:159-67.*
32. Pearl ML, Edgerton BW, Kazimiroff PA, Burchette RJ, Wong K. Arthroscopic release and latissimus dorsi transfer for shoulder internal rotation contractures and glenohumeral deformity secondary to brachial plexus birth palsy. *J Bone Joint Surg Am 2006;88-3:564-74.*
33. Sever JW. Obstetric paralysis: report of eleven hundred cases. *JAMA 1925;85:1862-5.*
34. Gilbert A, Brockman R, Carlioz H. Surgical treatment of brachial plexus birth palsy. *Clin Orthop Relat Res 1991-264:39-47.*
35. Gilbert A. Tendon transfers for shoulder paralysis in children. *Hand Clin 1988;4-4:633-42.*
36. Immerman I, Valencia H, DiTaranto P, DelSole EM, Glait S, Price AE, Grossman JA. Subscapularis slide correction of the shoulder internal rotation contracture after brachial plexus birth injury: technique and outcomes. *Tech Hand Up Extrem Surg 2013;17-1:52-6.*
37. Birch R, Chen L. The medial rotation contracture of the shoulder in obstetric brachial plexus palsy. *J Bone Joint Surg Br 1996;78-B-Supp 1:68.*
38. van der Sluijs JA, van Ouwerkerk WJ, de Gast A, Nollet F, Winters H, Wuisman PI. Treatment of internal rotation contracture of the shoulder in obstetric brachial plexus lesions by subscapular tendon lengthening and open reduction: early results and complications. *J Pediatr Orthop B 2004;13-3:218-24.*

39. L'Episcopo JB. Restoration of muscle balance in the treatment of obstetrical paralysis. *N Y J Med 1939;39:357-60.*
40. Hoffer MM, Wickenden R, Roper B. Brachial plexus birth palsies. Results of tendon transfers to the rotator cuff. *J Bone Joint Surg Am 1978;60-5:691-5.*
41. Ozturk K, Bulbul M, Demir BB, Buyukkurt CD, Ayanoglu S, Esenyel CZ. Reconstruction of shoulder abduction and external rotation with latissimus dorsi and teres major transfer in obstetric brachial plexus palsy. *Acta Orthop Traumatol Turc 2010;44-3:186-93.*
42. Waters PM, Bae DS. Effect of tendon transfers and extra-articular soft-tissue balancing on glenohumeral development in brachial plexus birth palsy. *J Bone Joint Surg Am 2005;87-2:320-5.*
43. Dodwell E, O'Callaghan J, Anthony A, Jellicoe P, Shah M, Curtis C, Clarke H, Hopyan S. Combined glenoid anteversion osteotomy and tendon transfers for brachial plexus birth palsy: early outcomes. *J Bone Joint Surg Am 2012;94-23:2145-52.*
44. Wickstrom J, Haslam ET, Hutchinson RH. The surgical management of residual deformities of the shoulder following birth injuries of the brachial plexus. *J Bone Joint Surg Am 1955;37-A-1:27-36; passim.*
45. Zancolli EA. Classification and management of the shoulder in birth palsy. *Orthop Clin North Am 1981;12-2:433-57.*
46. Birch R. BG, Wynn Parry, C. B. *Surgical disorders of the periferal nerves.* Chuchill Livingstone, 1998:229-30.
47. Pearl ML, Edgerton BW. Glenoid deformity secondary to brachial plexus birth palsy. *J Bone Joint Surg Am 1998;80-5:659-67.*
48. Waters PM, Smith GR, Jaramillo D. Glenohumeral deformity secondary to brachial plexus birth palsy. *J Bone Joint Surg Am 1998;80-5:668-77.*
49. Friedman RJ, Hawthorne KB, Genez BM. The use of computerized tomography in the measurement of glenoid version. *J Bone Joint Surg Am 1992;74-7:1032-7.*
50. van der Sluijs JA, van Ouwerkerk WJ, de Gast A, Wuisman PI, Nollet F, Manoliu RA. Deformities of the shoulder in infants younger than 12 months with an obstetric lesion of the brachial plexus. *J Bone Joint Surg Br 2001;83-4:551-5.*
51. Di Mascio L, Chin KF, Fox M, Sinisi M. Glenoplasty for complex shoulder subluxation and dislocation in children with obstetric brachial plexus palsy. *J Bone Joint Surg Br 2011;93-1:102-7.*
52. Pearl ML, Edgerton BW, Kon DS, Darakjian AB, Kosco AE, Kazimiroff PB, Burchette RJ. Comparison of arthroscopic findings with magnetic resonance imaging and arthrography in children with glenohumeral deformities secondary to brachial plexus birth palsy. *J Bone Joint Surg Am 2003;85-A-5:890-8.*
53. Hoffer MM, Phipps GJ. Closed reduction and tendon transfer for treatment of dislocation of the glenohumeral joint secondary to brachial plexus birth palsy. *J Bone Joint Surg Am 1998;80-7:997-1001.*
54. Zancolli A, Zancolli R. Reconstructive surgery in brachial plexus sequelae. In: Gupta A, Kay S, Scheker LR, eds. *The Growing Hand*. London: Mosby, 2000:805-23.
55. Poyhia TH, Nietosvaara YA, Remes VM, Kirjavainen MO, Peltonen JI, Lamminen AE. MRI of rotator cuff muscle atrophy in relation to glenohumeral joint incongruence in brachial plexus birth injury. *Pediatr Radiol 2005;35-4:402-9.*
56. Hogendoorn S, van Overvest KL, Watt I, Duijsens AH, Nelissen RG. Structural changes in muscle and glenohumeral joint deformity in neonatal brachial plexus palsy. *J Bone Joint Surg Am 2010;92-4:935-42.*

57. Waters PM, Monica JT, Earp BE, Zurakowski D, Bae DS. Correlation of radiographic muscle cross-sectional area with glenohumeral deformity in children with brachial plexus birth palsy. *J Bone Joint Surg Am 2009;91-10:2367-75.*
58. Weekley H, Nikolaou S, Hu L, Eismann E, Wylie C, Cornwall R. The effects of denervation, reinnervation, and muscle imbalance on functional muscle length and elbow flexion contracture following neonatal brachial plexus injury. *J Orthop Res 2012;30-8:1335-42.*
59. Nikolaou S, Peterson E, Kim A, Wylie C, Cornwall R. Impaired growth of denervated muscle contributes to contracture formation following neonatal brachial plexus injury. *J Bone Joint Surg Am 2011;93-5:461-70.*
60. Curtis C, Stephens D, Clarke HM, Andrews D. The active movement scale: an evaluative tool for infants with obstetrical brachial plexus palsy. *J Hand Surg Am 2002;27-3:470-8.*
61. Clarke HM, Curtis CG. An approach to obstetrical brachial plexus injuries. *Hand Clin 1995;11-4:563-80; discussion 80-1.*
62. Mallet J. [Obstetrical paralysis of the brachial plexus. II. Therapeutics. Treatment of sequelae. Priority for the treatment of the shoulder. Method for the expression of results]. *Rev Chir Orthop Reparatrice Appar Mot 1972;58:Suppl 1:166-8.*
63. Birch R, Bonney G, Dowell J, Hollingdale J. Iatrogenic injuries of peripheral nerves. *J Bone Joint Surg Br 1991;73-2:280-2.*
64. Bae DS, Waters PM, Zurakowski D. Reliability of three classification systems measuring active motion in brachial plexus birth palsy. *J Bone Joint Surg Am 2003;85-A-9:1733-8.*
65. Westin B HT, von Koch L. Obstetric brachial plexus injury: expectations before and satisfaction three months after secondary surgery on the shoulder. *Hand Therapy 2012;17-4:95-9.*
66. Langer JS, Sueoka SS, Wang AA. The importance of shoulder external rotation in activities of daily living: improving outcomes in traumatic brachial plexus palsy. *J Hand Surg Am 2012;37-7:1430-6.*
67. Ho ES, Curtis CG, Clarke HM. The brachial plexus outcome measure: development, internal consistency, and construct validity. *J Hand Ther 2012;25-4:406-16; quiz 17.*
68. Compston A. Aids to the investigation of peripheral nerve injuries. Medical Research Council: Nerve Injuries Research Committee. His Majesty's Stationery Office: 1942; pp. 48 (iii) and 74 figures and 7 diagrams; with aids to the examination of the peripheral nervous system. By Michael O'Brien for the Guarantors of Brain. Saunders Elsevier: 2010; pp. [8] 64 and 94 Figures. *Brain 2010;133-10:2838-44.*
69. Friden J, Lieber RL. Spastic muscle cells are shorter and stiffer than normal cells. *Muscle Nerve 2003;27-2:157-64.*
70. Lieber RL, Runesson E, Einarsson F, Friden J. Inferior mechanical properties of spastic muscle bundles due to hypertrophic but compromised extracellular matrix material. *Muscle Nerve 2003;28-4:464-71.*
71. Dubowitz V, Sewry CA. *Muscle Biopsy- A practical approach.* London: Bailliere Tindall, 1985.
72. Gueorguieva R, Krystal JH. Move over ANOVA: progress in analyzing repeated-measures data and its reflection in papers published in the Archives of General Psychiatry. *Arch Gen Psychiatry 2004;61-3:310-7.*
73. Kirkos JM, Kyrkos MJ, Kapetanos GA, Haritidis JH. Brachial plexus palsy secondary to birth injuries. *J Bone Joint Surg Br 2005;87-2:231-5.*